Anesthesia Management of a Patient with Kearn's - Sayre Syndrome

Kearn's – Sayre Sendromlu Hastanın Anestezi Yönetimi

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Özet

Kearn's Sayre Sendromu birden fazla sistemi ilgilendiren, nadiren görülen bir çeşit mitokondriyal myopatidir. Bu olgu sunumunda Kearn's Sayre Sendromlu hastanın açık kolesistektomi ve pyeloplasti ameliyatının kombine spinal epidural anestezi altında başarıyla gerçekleştirilmesi ve mitokondriyal myopatili hastaların anestezi yönetiminin planlanmasında dikkat edilmesi gereken noktaları belirtmeye çalıştık.

Anahtar Kelimeler

Kearn's Sayre Sendromu; Mitokondriyal Miyopati; Kombine Spinal Epidural Anestezi; Anestezi Yönetimi

Abstract

Kearn's Sayre Syndrome is a multi-system mitochondrial myopathy which has rarely seen. We report a patient with Kearn's Sayre syndrome who had laparoscopic cholesistectomy and pyeloplasty operation under combined spinal epidural anesthesia and importance of anesthesia management of this syndrome.

Keywords

Kearn's Sayre Syndrome; Mitochondrial Myopathy; Combined Spinal Epidural Anesthesia; Anesthesia Management

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Introduction

Kearn's Sayre syndrome (KSS) is one of the clinical presentations of mitochondrial myopathy. Symptoms of this syndrome include characteristic appearance before second decade with chronic progressive external ophthalmoplegia, retinal pigment degeneration and heart block, while it is occasionally combined with other neurologic and endocrinologic symptoms [1]. We report a patient with KSS who had underwent laparoscopic cholecystectomy and pyeloplasty operation under combined spinal epidural anesthesia (CSEA) with discussion of its clinical characteristics and anesthetic management.

Case Report

A 30- year –old female patient who suffered from right side pain, abdominal swelling, dizziness, general fatigue and swallowing difficulties was admitted to the emergency department of a tertiary hospital. Except for an AST value of 94 U/L, other laboratory parameters were unremarkable. Ultrasonography evaluation revealed bladder stones and right ureteropelvic obstruction. She was scheduled to have emergent laparoscopic cholecystectomy and pyeloplasty operation.

Her medical history included mitochondrial myopathy and compensated renal failure. She had initial muscle cramps, muscle weakness and exercise intolerance since she was 12 years old. The definitive diagnosis of mitochondrial myopathy (RRF(+) Cox – lipid increased) was made by skeletal muscle biopsy.

Physical examination revealed a short stature (149cm), cachexia and severe muscle wasting (body weight of 34 kg, BMI= 15). She had ptosis, limitation of movement in both eyes and chronic external ophtalmoplegia. Neurological examination included general muscle atrophy, hypoactive deep tendon reflexes and motor force of ¾ at four extremities. Occulopharyngeal dystrophy caused swallowing difficulties and ear nose throat examination showed palatum mole paralysis.

On arrival in the operating room, oxygen was applied at a rate of 2 L/minute through a nasal cannula. An intravenous preload of 500 mL of 5% dextrose solution was given over 30 minutes and was followed by IV infusion of 0.9% serum physiologic solution at a rate of 5 mL/kg/h during the operation. Standard monitoring of electrocardiogram, blood pressure, pulse oxymetry and temperature were applied. In addition, arterial catheterization was considered to facilitate continuous blood pressure monitoring and frequent sampling for blood glucose, blood gases and serum lactate levels. Temperature monitoring with a nasopharyngeal probe was essential to maintain normothermia. CSEA was performed at the L2-L3 interspace with awake patient at lateral position using needle-through-needle technique via a single interspace (Espocan, B. Braun, Melsungen, Germany). The procedure included an initial application of 18-G Tuohy epidural needle (Perican 88 mm), followed by performance of spinal block with administration of 12.5 mg 0.5% hyperbaric bupivacaine via a 27-G spinal needle (Spinocan 25 mm). The adequacy of the sensory block was assessed with pinprick test and motor block with Bromage scale. Sensory block was achieved up to T4 dermatome and patient was turned to supine position. There was no requirement for additional local anesthetic administration through epidural catheter during the operation. Arterial blood pressure was well maintained during the operation without any vasoactive drug support. Blood glucose, arterial blood gases, serum lactate levels, temperature were in the normal range during the operation.

After surgery, the patient was equipped with a patient controlled analgesia (PCA) device. The analgesic mixture containing 2 mg/mL of bupivacaine and 2 μg/mL of fentanyl mixture was prepared. Demand dose was 5 mL, background infusion was 5 mL/h, and lockout interval was 30 minutes. Rescue analgesic and sedative drugs were not required during the operation and in the postoperative period. The postoperative period transpired without adverse events and the patient was discharged 24-hour after surgery.

Discussion

KSS is a kind of mitochondrial myopathy which is rarely seen. It is caused by mitochondrial deoxyribonucleic acid deletion. Mitochondrial myopathies are a heterogeneous group of disorders of muscle energy metabolism [1, 2].

During anesthesia management of patients with mitochondrial myopathies, we must sustain normothermia, normoglycemia as well as avoid metabolic stress in order to preserve energy production by the diseased mitochondria.

Preoperative fasting is a kind of metabolic stress for the patients with mitochondrial myopathies. Intravenous fluids should be initiated during the preoperative fasting period to allow maintenance of normoglycemia, as excessive glycolytic oxidation of glucose in this patient group may increase plasma lactate levels. Lactate free IV fluids were recommended (such as 5% dextrose–0.9 % saline) [3].

We must be aware of "full stomach" precautions while giving anesthesia to this kind of patients. Our patient had palatum molle paralysis which might predispose to aspiration of gastric contents, so preferring regional anesthesia technique was an advantage to avoid aspiration risk [2].

Low blood glucose level should be avoided. However, high blood glucose may indicate an acute disturbance in pyruvate metabolism or oxidative phosphorylation. We measured blood glucose levels pre- and postoperatively so that no problem was encountered.

We sustained normothermia during operation and in the postoperative period. It is essential to maintain body temperature during perioperative period because hypothermia is known to depress mitochondrial function and trying to regain normothermia after surgery causes metabolic stress. Because of this reason, temperature should be measured in patients having major surgery under anesthesia [4].

Mitochondria are a potential site of action of general and local anaesthetics. As the central nervous system has a high demand for energy, patients with mitochondrial myopathies may be sensitive to anaesthesia [5]. Literature shows that both general and regional anesthesia have been used successfully [1-5]. Some anesthetic considerations have been proposed. It is very important to maintain adequate oxygen balance, stable cardiovascular function, and good gas exchange. The anesthetic plan for patients with mitochondrial myopathy should routinely include malignant hyperthermia precautions. Patients with mitochondrial myopathies were predisposed to malignant hyperthermia or myasthenia like sensitivity to neuromuscular blockade [1-4]

The use of regional anaesthesia eliminates the risk of prolonged muscle relaxation, central nervous system depression and the possibility of malignant hyperthermia. Aerobic metabolism is dysfunctional at mitochondrial myopathy patients. Regional anesthesia also provides adequate surgical anesthesia and good postoperative analgesia and also prevents the increases in oxygen consumption and metabolic rate [1,5,6]

Skeletal muscle weakness may compromise postoperative ven-

tilation, especially after upper abdominal or thoracic surgery. Our patient had spontaneous breathing during operation under spinal anesthesia and we did not have any necessity to use opioids or sedatives during operation. If it was necessary, all opioids and sedative-hypnotics should be titrated carefully due to the decreased ventilatory response to hypoxia and hypercarbia, particularly as opioids may further impair regulation of breathing and may lead to a respiratory acidosis [7].

At KSS, heart rhythm problems, specifically severe conduction blocks may appear which can lead to death. Dilated cardiomyopathy and congestive heart failure may develop. Therefore, fluid overload should be avoided in the perioperative period. The deletion of mitochondrial deoxyribonucleic acid in the myocardium may be the cause of myocardial dysfunction [8]. Our patient did not have any heart rhythm problems in the perioperative period. .The patient was hemodynamically stable throughout the operation. She was comfortable, her vital signs were stable and she was subsequently discharged.

In summary, we report a patient with KSS who underwent laparoscopic cholecystectomy and pyeloplasty operation successfully under combined spinal epidural anesthesia. By using the appropriate anesthesia management of mitochondrial myopathies (KSS), we can reduce the risks of anesthesia.

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